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Consensus Statement of the Movement Disorder Society on Tremor

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Summary: This is a proposal of the Movement Disorder Society for a clinical classification of tremors. The classification is based on the distinction between rest, postural, simple kinetic, and intention tremor (tremor during target-directed movements). Additional data from a medical history and the results of a neurologic examination can be combined into one of the following clinical syndromes defined in this statement: enhanced physiologic tremor, classical essential tremor (ET), primary orthostatic tremor, task- and position-specific tremors, dystonic tremor, tremor in Parkinson's disease (PD), cerebellar tremor, Holmes' tremor, palatal tremor, drug-induced and toxic tremor, tremor in peripheral neuropathies, or psychogenic tremor. Conditions such as asterixis, epilepsia partialis continua, clonus, and rhythmic myoclonus can be misinterpreted as tremor. The features distinguishing these conditions from tremor are described. Controversial issues are outlined in a comment section for each item and thus reflect the open questions that at present cannot be answered on a scientific basis. We hope that this statement provides a basis for better communication among clinicians working in the field and stimulates tremor research. **Key Words:** Tremor—Classification—Clinical assessment.

The present effort has been undertaken to create a common terminology for tremor disorders within the Movement Disorder Society (MDS). The article was discussed during the symposium on tremor in Kiel, Germany, subsequently distributed within the Scientific Issues Committee of the MDS, and finally approved by the EXCO of the MDS. One goal of the MDS is to foster such consensus statements. We hope that this article will be the first of several to be published on different issues.

Authorities in the field of tremor differ in their views of tremors because similar observations are interpreted differently. It cannot be the intent of this article to solve all of these controversies because scientific data are lacking for many of them. However, it is our goal to summarize the minimal consensus, to address the controversial issues, and to provide a framework in which unresolved questions can be addressed.

Tremor can be classified in different ways. Because of the numerous etiologies for tremor, a practical etiologic classification or a valid physiologic classification is not available. Therefore, the gold standard for tremor research remains clinical classification. We propose to classify tremors according to the traditional approach of clinical neurology. Based on their specific clinical features, tremors are grouped into syndromes that can be separated on the basis of clinical observations alone. This article is based on the assumption that different components of tremor can be separated by clinical observation and need to be described with a common terminology (see 1.1, Definitions of the Phenomenology of Tremor). To achieve this objective, the meaning and interpretation of some tremor-related tests must be defined (see 1.2,

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Definitions of Terms Related to Tremor or Tremulous Movements). The results of the neurologic examination should be described in a generally accepted form (see 1.3. Clinical Assessment of Tremor Patients). The diagnostic classification consists of data from the history and medical examination of a patient, which is sufficient to classify a specific tremor among a number of discrete tremor syndromes (see 2.1, Syndromic Classification of Tremor).

This classification is based on earlier work by members of the MDS [1–3] and we would not have been able to finish this statement without the help of all the colleagues mentioned in the Acknowledgments.

Some issues have been discussed at length within the scientific committee (for example, the definition of rest and intention tremor). The latter in particular has been revived because some recent articles tended to lump this feature of high diagnostic value with other kinetic tremors. The coactivation sign of psychogenic tremor is now included among the diagnostic tests for tremors.

This statement defines tremor syndromes on clinical grounds that have not been detailed before or have been defined in conflicting ways in the past. Furthermore, new names for tremors were introduced. The statement provides a clinical definition of enhanced physiologic tremor. A subclassification of essential tremor (ET) and of parkinsonian tremor is provided. Dystonic tremor is considered now as a separate entity. Palatal tremor is now classified among the tremors and Holmes' tremor has been introduced as a substitute for the misnomer, rubral tremor.

It is the intent of this consensus statement to stimulate discussions within and outside the MDS and thus to contribute to the development of tremor research.

1. PHENOMENOLOGY OF TREMOR

1.1 Definitions referring to the phenomenology of tremor

Critical to clinical analysis and classification of tremor is a definition of the activation conditions during which tremor occurs. The following definitions and related criteria are based on the Tremor Investigation Group (TRIG) criteria[3] and cover the terms that are, in our view, necessary and sufficient to describe the clinical spectrum of tremors (Table 1).

1.1.1. Tremor

Rhythmical, involuntary oscillatory movement of a body part

Comment: The amplitude of tremor is not critical to the definition. Small-amplitude tremors may only be detectable by sensitive recording devices. In psychogenic tremor, the tremor may be produced voluntarily, although awareness that the tremor is voluntary may be subconscious.

1.1.2. Tremor envelope

This is the profile of the visible tremor amplitude during movement, for example, the finger-to-nose test.

1.1.3. Rest tremor

Rest tremor is defined by tremor that occurs in a body part that is not voluntarily activated and is completely supported against gravity (ideally, resting on a couch).

Comment: This definition was a matter of debate and is a minimal consensus as it stands now. The major scientific problem was to exclude postural tremors that continue when the limb is supported, which sometimes occurs in ET, dystonic tremor, psychogenic tremors, and other conditions. Some of us believe that it is based on a subconscious or conscious ongoing contraction of the respective limb even when the limb is supported.

Most experts agree that a practical way to separate typical rest tremor from action tremor persisting during rest conditions is to investigate the patient during target-directed movements. Tremor amplitude almost always diminishes during these movements in rest tremor. In severe action tremors that persist also under rest conditions, increasing or constant tremor amplitudes are found during voluntary movements.

For rest tremor, the tremor amplitude increases during mental stress (counting backwards, stroop test, and so forth) or when movements of another body part are performed (especially walking). Although tremors may be aggravated by mental load, increasing tremor amplitudes are not seen in other tremors when the patient is lying on a couch, completely relaxed during mental stress.

Rest tremor is a physiologically and clinically separate entity probably generated by mechanisms unique to this symptom. It is mostly found in Parkinson's disease (PD) but seldom in other conditions. According to clinical experience, rest tremor responds in most patients to dopaminergic treatment. However, because exceptions exist and sufficient studies on this question were not available, it was discarded from the definition.

1.1.4. Action tremor

Action tremor is any tremor that is produced by voluntary contraction of muscle, including postural, isometric, and kinetic tremor. The latter includes intention tremor.

1.1.4.1. Postural tremor

Postural tremor is present while voluntarily maintaining a position against gravity.

Comment: This assumes voluntary or purposive activation of muscle necessary for the maintenance of position. It is recognized that postural tremor may appear or become exacerbated in specific (usually visually guided) postures. This type of tremor may be called position-specific or position-sensitive postural tremor.

1.1.4.2. Kinetic tremor

Tremor occurring during any voluntary movement.

Comment: Kinetic tremor may occur in nonvisually guided and visually guided movements (it does not matter if a nongoal-directed movement like repetitive flexion/extension movements of the hands or a goal-directed movement [for example, during the finger-to-nose test] is considered).

1.1.4.2.1. Simple kinetic tremor

This tremor occurs during voluntary movements that are not target-directed.

Comment: The term, simple kinetic tremor, was chosen to separate nontarget-directed from target-directed tremors. For example, it is tested with simple pronation/supination movements or flexion/extension wrist movements. Simple kinetic tremor may persist during voluntary, goal-directed movement without an increase in amplitude during the terminal phase of movement. Some patients have larger amplitudes during particular trajectories, postures, or movements.[4]

1.1.4.2.2. Tremor during target-directed movements (*intention tremor*)

Classic intention tremor is present when amplitude increases during visually guided movements toward a target at the termination of the movement[5] and the possibility of a position-specific tremor or a postural tremor produced at the beginning or end of a movement is excluded.

Comment: Whenever a tremor increases substantially during the pursuit of a target or goal, it can be inferred that a disturbance of the cerebellum and its afferent or efferent pathways is present.[2,6–8] Typically, tremor amplitude fluctuates significantly as the target is approached, sometimes even from beat to beat. Although this feature is characteristic for intention tremor, it is not thought to be required for the definition as long as the possibility of a position-specific tremor at the end of a movement is excluded.

Intention tremor has sometimes been confused with action myoclonus and can be difficult to distinguish from ataxia in some patients. As long as the dominant feature of the movement disorder is rhythmicity, it should be labeled as tremor.

Use of the term, intention tremor, has often been criticized because it is not the mental intention that aggravates the tremor but the performance of target-directed, especially visually guided, movements. However, because the term is ingrained in the neurologic culture, we kept it but defined it as mentioned previously.

1.1.4.3. Task-specific kinetic tremor

Kinetic tremor may appear or become exacerbated during specific activities. Occupational tremors and primary writing tremor are examples of this kind of tremor.

1.1.4.4. Isometric tremor

Tremor occurring as a result of muscle contraction against a rigid stationary object (for example, while making a fist or squeezing the examiner's fingers).

Comment: This form of force tremor can occur in isolation or with other tremor symptoms and may be the cause of separate complaints.

1.2 Definitions of terms related to tremor or tremulous movements

1.2.1. Tone

The resistance encountered by the examiner when a limb or other body part is moved passively about a joint in the relaxed state.

1.2.2. Reinforced tone

An increase in resistance during passive movement about a joint induced by voluntary activity in a noncontiguous body segment.

1.2.3. Rigidity

Increased tone within the range of passive movement about a joint.

1.2.4. Cogwheel phenomenon

Rhythmic brief increase in resistance during passive movement about a joint.

1.2.5. Froment's (muscle tone) sign

Increase in resistance to passive movements of a limb about a joint that can be detected specifically when there is a voluntary activity of another body part.

Comment: This phenomenon may be seen in a wide variety of tremulous disorders including ET[9] and PD.[10] The definition of Froment's sign adapted earlier is different from the original description[11] but has become the most common practical method of eliciting this sign.

1.2.6. Coactivation sign of psychogenic tremor

Resistance to passive movements about a joint (tested for rigidity) in a trembling limb with the following char-

acteristics[12]: the occurrence of tremor is dependent on the occurrence of an increase in the tone of the respective limb (that is, the examiner often feels the patient "fighting" against her or him); and the rhythmic resistance (tremor) disappears with the voluntarily increased tone. Additionally, the frequency may change during this maneuver.

Comment: Cogwheeling in the setting of PD or ET differs from the present coactivation sign because the latter disappears when the patient completely relaxes. Coactivation in psychogenic tremor resembles voluntary co-contraction with overlying trembling.[13] In contrast to the cogwheel rigidity of PD, the muscles can be completely relaxed with appropriate challenge. Additionally, coactivation may sometimes produce some abnormal (or even pseudodystonic) positioning of the hands during tremor.

This feature is often seen in psychogenic tremor and may be related to a clonus mechanism underlying psychogenic tremor.[12] When the voluntary or pseudovoluntary coactivation disappears, the tremor disappears also. However, this sign is not seen in all the patients with psychogenic tremor because other mechanisms (like voluntary shaking) may be operating.

1.2.7. Dystonia

A syndrome dominated by sustained muscle contractions, frequently causing twisting and repetitive movement or abnormal postures.

1.3 Clinical assessment of tremor patients

A common understanding of a movement disorder needs a common language. The following outline may be helpful in communicating the examination of tremor phenomenology.

1.3.1 Inspection of the tremor

Topography

Table 1 lists the parts of the body that can be clinically and meaningfully separated when describing tremor topography.

Activation condition of tremor (For definitions, see 1.1.)

Table 2 shows the different terms for the description of tremors according to the activation conditions.

Frequency

- Low (<4 Hz)
- Medium (4–7 Hz)
- High (>7 Hz)

TABLE 1. *Parts of the body that have been described to be affected by tremor*

Head	Trunk
Chin	Lower extremity
Face	Hip
Tongue	Knee
Palate	Ankle joint
Upper extremity	Toes
Shoulder	
Elbow	
Wrist	
Fingers	

1.3.2 Specific examinations for assessment of:

- Akinesia/bradykinesia
- Muscle tone (including Froment's sign for the upper and lower extremity and coactivation sign for psychogenic tremor)
- Postural abnormalities
- Dystonia
- Cerebellar signs
- Pyramidal signs
- Neuropathic signs
- Systemic signs (thyrotoxicosis and so forth)
- Gait and stance (orthostatic tremor)

1.3.3 Specific data from the medical history

- Onset of tremor
- Family history of neurologic diseases (especially tremor)
- Alcohol sensitivity
- Sequence of spread of tremor (including symmetry and laterality) associated diseases (restless legs, polyneuropathy, and so forth)
- Medication
- Drug abuse

1.4 Rating scales for tremors

Several scales have been proposed; in principle, it is useful to distinguish between rest tremors and action

TABLE 2. *Terminology for tremor and the hierarchical relation of the terms as indicated by the numbers*

1.	Rest tremor
2.	Action tremor
2.1	Postural tremor
2.1.1	Position-independent postural tremor
2.1.2	Position-specific postural tremor
2.2	Kinetic tremor
2.2.1	Unspecified kinetic tremor (non-goal-directed movements, for example, repetitive flexion-extension movements)
2.2.2	Tremor during goal-directed movements (intention tremor)
2.2.3	Task-specific kinetic tremor
2.2.4	Isometric tremor

tremors, which are associated with different symptoms; the scale should also avoid mixing measures of impairment with disability or handicap. Moreover, difficulties arise from the variability of tremor. For rest tremors, the Unified Parkinson's Disease Rating Scale (UPDRS)[14] with some supplementary questions[15] has been proposed. For postural and intention tremor, two different scales have been proposed.[16–19]

2.1 Syndromic classification of tremor

The phenomenology of tremor elements can be combined into some specific syndromes. They are the basis for further diagnostic and therapeutic procedures (Table 3).

2.1.1. Physiologic tremor

Physiologic tremor is present in every normal subject and every joint or muscle that is free to oscillate. Normal finger tremor can just be seen with the naked eye.

Comment: The amplitude of physiologic tremor is low; its frequency is high for the hand and fingers but low for proximal joints. Mechanical and neural factors cause this tremor.[20–22]

2.1.2. Enhanced physiologic tremor syndrome

This syndrome is defined by both of the following:

1. Easy visibility of the tremor, mainly postural and high-frequency.
2. No evidence of an underlying neurologic disease; the cause of tremor is usually reversible.

Comment: This definition covers many tremor etiologies, typically those elicited by endogenous or exogenous intoxication. These tremors are mostly reversible, provided that the cause of tremor is identified and corrected. Once such a syndrome has been identified, the etiology needs to be clarified.

These criteria provide a clinical definition for a term that has been traditionally used as a condition defined by physiologic measurements.[23–25] Nevertheless, this definition covers all those tremors (and more) that have been described as conditions of enhanced physiologic tremor.[20,25] The important mechanisms underlying these tremors are a mechanical component activated by a stretch reflex component. However, alternatively (or additionally), this tremor can also be caused by an enhanced central tremor oscillation that is also seen in normal subjects and has been labeled the 8–12 Hz central tremor component.[22,26,27]

2.1.3. Essential tremor

Comment: ET has acquired two meanings over recent years. On one hand, it is a tremor syndrome classically defined by a mostly hereditary, mainly postural tremor of the hands and sometimes of the head.[9,28,29] ET is the most common movement disorder and accepted criteria are necessary, especially for epidemiologic studies. On the other hand, there are tremors of unknown origin (so-called idiopathic or essential), but with distinct clinical symptoms that usually distinguish these tremors on clinical grounds from the classic ET. To include these two entities in a unique classification, the TRIG has proposed definite and probable ET as the term for the so-called classic ET and possible ET as the term for the remaining entities,[30] which are modified here.

2.1.3.1. Classic ET

For the classic ET described and elaborated in many studies,[2,9,28,29,31–58] the following definition seems sufficient:

Inclusion criteria

1. Bilateral, largely symmetric postural or kinetic tremor involving hands and forearms that is visible and persistent.
2. Additional or isolated tremor of the head may occur but in the absence of abnormal posturing.

Exclusion criteria

1. Other abnormal neurologic signs, especially dystonia.
2. The presence of known causes of enhanced physiologic tremor, including current or recent exposure to tremorogenic drugs or the presence of a drug withdrawal state.
3. Historic or clinical evidence of psychogenic tremor.
4. Convincing evidence of sudden onset or evidence of stepwise deterioration.
5. Primary orthostatic tremor.
6. Isolated voice tremor.
7. Isolated position-specific or task-specific tremors, including occupational tremors and primary writing tremor.
8. Isolated tongue or chin tremor.
9. Isolated leg tremor.

Comment: This definition includes the TRIG criteria for definite and probable ET. However, in our view, tremor duration is no longer a criterion because the 3-year (probable ET) and 5-year (definite ET) duration criteria were arbitrary. In kindreds with hereditary ET, younger members of the family may have ET for a shorter duration. Similarly, marked ET can occur in

TABLE 3. *Syndromic classification of tremor*

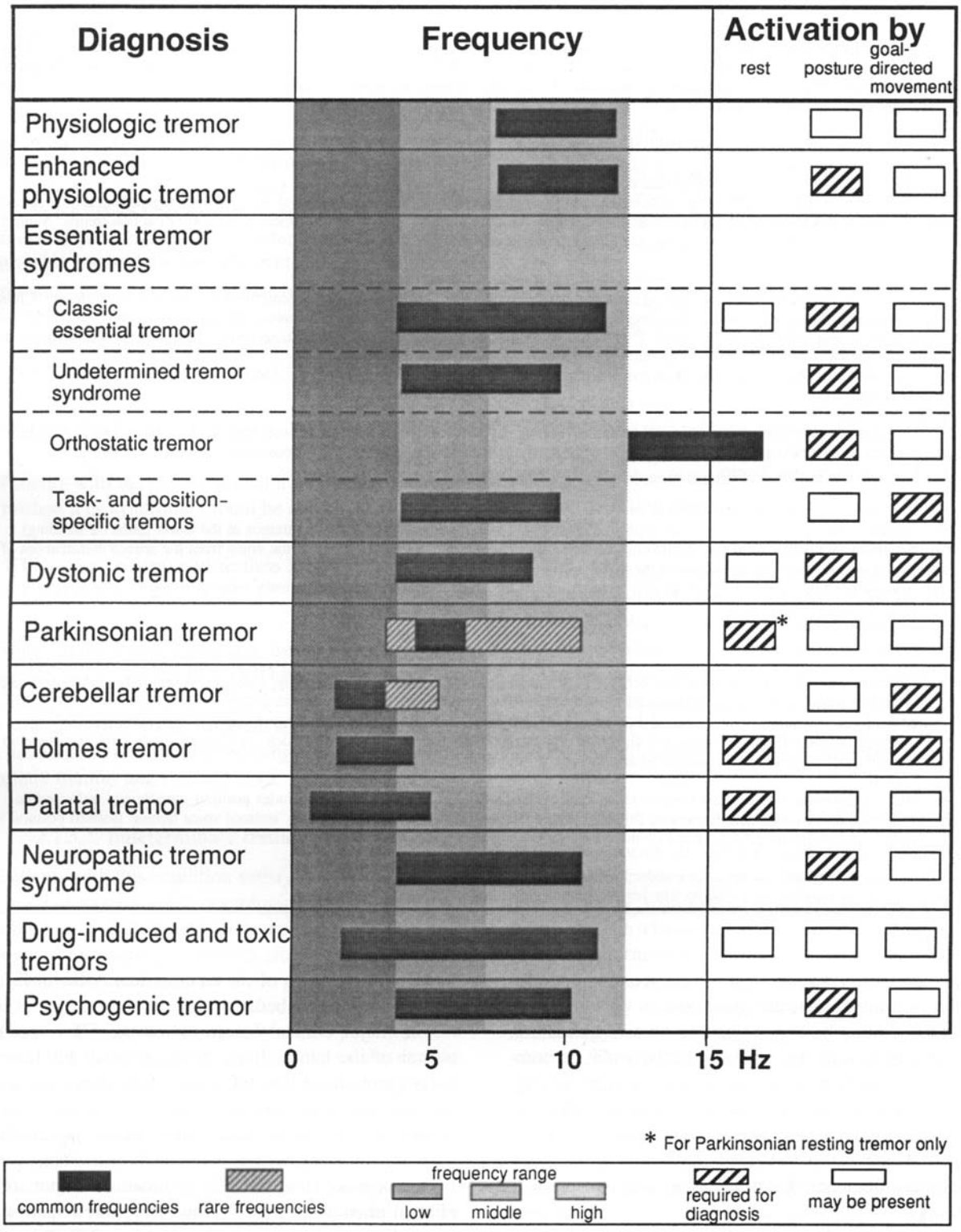

TABLE 4. *TRIG classification of essential tremor*[30]

Essential tremor refers to a phenomenological criterion. It is not a function of disability, pathophysiology, or heredity.

A. Definite essential tremor
   1. Inclusion criteria
      (i) Tremor: Bilateral postural tremor with or without kinetic tremor, involving hands and forearms, that is visible and persistent. (Tremor of other body parts may be present in addition to upper limb tremor. Bilateral tremor may be asymmetric. Tremor is reported by patient to be persistent, although the amplitude may fluctuate. Tremor may or may not produce disability.)
      (ii) Duration: Longer than 5 years
   2. Exclusion criteria
      (i) Other abnormal neurologic signs (with the exception of the presence of tremor and Froment's sign, the full neurologic examination should be normal for age)
      (ii) Presence of known causes of enhanced physiologic tremor
      (iii) Concurrent or recent exposure to tremorogenic drugs or the presence of a drug withdrawal state (Many drugs acting on the central nervous system can produce tremor as a side effect. In people, drug-induced tremor is most often in the form of action tremor. Subjects should be drug-free for a period exceeding the known biologic effect of the drug.)
      (iv) Direct or indirect trauma to the nervous system within 3 months preceding the onset of tremor (This includes head injury [direct or indirect], and peripheral injury, if the anatomic distribution is the same as that of the tremor.)
      (v) Historic or clinical evidence of psychogenic origins of tremor. (The definition of psychogenic tremor is itself open to debate. Clinical features that may suggest this are unphysiological variations [>1 Hz] in tremor frequency, unusual and inconsistent behavioral characteristics, and spontaneous remission. Psychiatric or social factors [multiple somatization, secondary gain, litigation or compensation pending] may support the diagnosis of psychogenic tremor.)
      (vi) Convincing evidence of sudden onset or evidence of stepwise deterioration

B. Probable essential tremor
   1. Inclusion criteria
      (i) The same as those for definite essential tremor. (Tremor may be confined to body parts other than hands. These may include head and postural tremor of the legs. However, abnormal posture of the head would suggest the presence of dystonic head tremor.)
      (ii) Duration longer than 3 years
   2. Exclusion criteria
      (i) The same as for definite essential tremor
      (ii) Primary orthostatic tremor (isolated, high-frequency [14–18 Hz] bilaterally synchronous tremor of the lower limbs on standing)
      (iii) Isolated voice tremor (because of the clinical difficulty of separating essential tremor of the voice from the speech disturbances of laryngeal dystonia and other dystonias of the vocal apparatus)
      (iv) Isolated position-specific or task-specific tremors, including occupational tremors and primary writing tremor
      (v) Isolated tongue or chin tremor

C. Possible essential tremor
   1. Inclusions
      (i) Type I
         a. Subjects who satisfy the criteria of definite or probable essential tremor but exhibit other recognizable neurologic disorders, such as parkinsonism, dystonia, myoclonus, peripheral neuropathy, or restless leg syndrome
         b. Subjects who satisfy the criteria of definite or probable essential tremor but exhibit other neurologic signs of uncertain significance not sufficient to make the diagnosis of a recognizable neurologic disorder. Such signs may include mild extrapyramidal features, such as hypomimia, decreased arm swing, or mild bradykinesia.
      (ii) Type II
         Monosymptomatic and isolated tremors of uncertain relation to essential tremor. This includes position-specific and task-specific tremors, such as occupational tremors, primary writing tremor, primary orthostatic tremor, isolated voice tremor, isolated postural leg tremor, and unilateral postural hand tremor.
   2. Exclusions
      The exclusions are the same as items 2–4 under Definite essential tremor.
      A further form of classification could be whether the tremor is familial or presumed sporadic.

middle age within 3 years of its onset, thus the neurologist has no doubt about the diagnosis.

Tremor of other body parts may be present in addition to upper limb tremor. The majority of the scientific committee classifies pure head tremor without signs of dystonia as classic ET, but some would not do this. Bilateral tremor may be asymmetric but not markedly so. Tremor is reported by patients to be persistent, although the amplitude may fluctuate. Tremor may or may not produce disability.

This definition does not refer to the severity of the tremor as earlier classifications did,[2,20] and the semiologic characteristics of the tremors that occur in ET are not precisely described.

The major clinical feature of classic ET is postural tremor of the hands. Early descriptions of ET have always emphasized that ET is not made strikingly worse during visually guided (target-directed) movements.[1,2,20,29,34] In the past, other clinical presentations with intention tremor (or even with rest tremor) have been recognized and partially included as variants of the clinical phenomenology.[20] However, more recent series show that intention tremor is not rare in ET. Therefore, an alternative definition of the clinical spectrum of hand

tremor in classic ET could be mainly postural tremor, but in many cases is intention tremor[20,27,36] and rarely, even tremor at rest occurs.[25,27,41,45,59–61] In other words, the presence of one specific condition that activates the tremor amplitude cannot exclude classic ET. The TRIG classification in its original form is included (see Table 4) because it may still be used for special epidemiologic purposes.

The category of possible ET defined by the TRIG has been reclassified; patients who were defined by TRIG as possible ET, Type I because they satisfied the criteria of definite or probable ET but exhibit other recognizable neurologic disorders (such as parkinsonism, dystonia, or peripheral neuropathy) have been respectively reclassified under:

1. Parkinsonian tremor syndromes
2. Dystonic tremor syndromes
3. Neuropathic tremor syndromes

Patients with definite or probable ET and myoclonus or restless legs syndrome should be described as having:

1. ET (as appropriate) and restless legs syndrome or
2. ET and myoclonus

Some monosymptomatic and isolated tremors have originally been classified by TRIG as possible ET, Type II. They have subsequently been better defined and thus are now classified among different subcategories, such as occupational tremor, primary writing tremor, primary orthostatic tremor, and isolated voice tremor (see later).

2.1.3.2. Indeterminate tremor syndrome

Patients with this condition satisfy the criteria of classic ET but exhibit other neurologic signs of uncertain significance not sufficient to make the diagnosis of a recognizable neurologic disorder (for example, mild extrapyramidal features, such as hypomimia, decreased arm swing, or mild bradykinesia).

Comment: In clinical practice when the patient is seen for the first time, it can be impossible to classify the condition definitely because the significance of the associated neurologic signs is not yet fully apparent. By labeling these patients with an indeterminate syndrome, the problems associated with an incorrect diagnosis or conflicting diagnoses is avoided and the neurologist remains open minded about therapeutic options. For example, distinguishing between early parkinsonism, ET, and especially, dystonic tremor, can be almost impossible in some elderly patients but the differentiation may become clearer with time. This category includes patients who were classified by TRIG as possible ET, Type Ib.

2.1.3.3. Primary orthostatic tremor

Orthostatic tremor is a unique tremor syndrome[62,63] characterized by:

1. A subjective feeling of unsteadiness during stance but only in severe cases during gait; patients rarely fall. None of the patients have problems when sitting and lying.
2. Sparse clinical findings that are mostly limited to a visible and occasionally, only palpable fine amplitude rippling of the leg (quadriceps or gastrocnemius) muscles when standing.
3. The diagnosis that can be confirmed only by EMG recordings (for example, from the quadriceps muscle) with a typical 13–18-Hz pattern. All of the leg, trunk, and even arm muscles can show this tremor, which is typically absent during tonic activation while the patient is sitting and lying.[64–68]

Comments: Falls have been observed mostly in elderly people, who may have additional neurologic problems (PD, senile gait disturbances, and so forth). Falls are obviously not a problem during the onset of the condition, which usually occurs during middle age.

The diagnosis critically depends on electromyographic (EMG) confirmation of the high-frequency EMG pattern because other tremors or symptoms (for example, akathisia, cerebellar stance tremor) during stance can occur with similar complaints.

2.1.3.4. Task and position-specific tremors

Several forms of tremor have been identified that share specific situations of activation as a common feature. Primary writing tremor is the most frequent example of such a tremor and occurs in specialized fields of motor performance. Other examples include specific tremors in musicians[69] and athletes.[70,71] These tremors can be clearly classified by means of their clinical symptomatology, which justifies their subclassification in this chapter. Unfortunately, this does not mean that these tremors can be unequivocally classified by means of their pathogenesis. It is still unknown whether these task-specific tremors represent a type of ET, a variant of dystonia, or a third, distinct pathophysiologic entity.

Comment: Isolated chin tremor was classified earlier among the tremors but it was generally thought that it should no longer be included here. Isolated chin tremor is an autosomal-dominant hereditary syndrome charac-

terized by a high-frequency, partially arrhythmic contraction of the mentalis muscles.[72,73] The onset is typically in infancy or childhood; geniospasm is the term that has been used. Recently, the question was raised as to whether or not this syndrome is a result of rhythmic myoclonus.[73] In the future, it may turn out to be the result of channelopathy.

2.1.3.4.1 Primary writing tremor

Primary writing tremor is present when tremor occurs only (or predominantly) during writing but not during other tasks in the active hand.

Comment: After the initial description,[74] writing tremor was extensively studied clinically.[49,75–89] Two forms of primary writing tremor have been described. Task-induced tremor is characterized by tremor appearing during writing only (Type A, task-specific tremor). If it occurs when the hand adopts a writing position, the terms position-specific or position-sensitive tremor (Type B) have been used.[87]

Some patients with tremor during writing also have dystonic posturing, and some patients with writer's cramp have tremor. The present definition excludes these so-called dystonic writing tremors and defines primary writing tremor as nondystonic. Dystonic writing tremors are presently covered among the dystonic tremors. However, it is still uncertain whether writer's tremor is a focal form (*forme fruste*) of an ET[77,78,80,87] or a dystonic tremor variant.[81–83,89,90] It may turn out that both possible etiologies are still covered under the present term of primary writing tremor and a reclassification may be necessary in the future.

2.1.3.4.2. Isolated voice tremor

Isolated voice tremor is present if vocalization is tremulous but no other parts of the body show tremor.

Comment: Isolated voice tremor[91] occurs in two variants. The first is often considered as a form of focal dystonia of the vocal cords[92–96]; the other is considered to be a variant of ET.[9,40,90,97,98]

Voice tremor does occur in other conditions (such as cerebellar disease or classic ET) together with additional tremor manifestations. However, to our knowledge, isolated voice tremor is limited to the previously mentioned conditions. Unfortunately, not much is known about these conditions, so the following provisional definition has been included only under the comments section:

The clinical diagnosis of isolated vocal tremor can be put forward if:

1. The tremor is limited to the voice.
2. Dystonic voice tremor is the more likely diagnosis (if the tremor ceases during emotional speech production (*gestes* maneuvers), singing, or changes in pitch.

2.1.4. The dystonic tremor syndromes

Dystonic tremor syndrome is still under debate and different definitions have been proposed by various clinicians.[40,69,74,76,77,82,83,99–108] We propose the following definitions:

2.1.4.1. Dystonic tremor

Tremor in a body part affected by dystonia.

1. Tremor in an extremity or body part that is affected by dystonia.
2. Focal tremors, usually with irregular amplitudes and variable frequency (mainly less than 7 Hz).
3. Mainly postural/kinetic tremors and usually not seen during complete rest.

Comment: A typical example of dystonic tremor is tremulous spasmodic torticollis (or dystonic head tremor). In many patients with dystonic tremor *gestes antagonistes* lead to a reduction of the tremor amplitude. This is especially true for dystonic tremor in cervical dystonia,[104,105] whereas essential head tremor does not show this sign. Tremors in body parts not affected by dystonia are excluded in this definition (see later).

2.1.4.2. Tremor associated with dystonia

This tremor occurs in a body part not affected by dystonia, but the patient has dystonia elsewhere.

Comment: For example, patients with cervical dystonia often show upper limb postural tremor[80,99,101–103,106,109,110] that is indistinguishable from enhanced physiologic tremor[111] or ET. Dystonic tremor and tremor associated with dystonia are different because postural tremors resembling mild classic ET can occur at extremities not involved in dystonia (for example, hand tremor in patients with otherwise monosymptomatic blepharospasm).

2.1.4.3. Dystonia gene-associated tremor

Tremor as an isolated finding in patients with a dystonic pedigree.

Comment: For example, isolated head tremor occurring in a patient with first-degree relatives with spasmodic torticollis.

2.1.5. Parkinsonian tremor syndromes

Tremor in PD is assumed if:

1. The patient has any form of pathologic tremor.
2. The patient has PD according to the brain bank criteria.[112] Bradykinesia must be present.

Comment: As a result of the variability of the clinical expression of tremors in PD, the definition is based on the general diagnosis of PD rather than on specific features of the tremors. Only the rest tremor component is, by itself, a positive diagnostic criterion for PD but other tremors are often seen in PD.

Different clinical presentations of tremors can be found in parkinsonian syndromes and are classified according to their clinical features. We decided to label them according to the state of activation of the limb when tremor is present. Pure rest tremor is infrequent; more common is the combination of rest and postural/kinetic tremors, which occurs in two variants.[10,113,114] Compared with those, isolated postural/kinetic tremors are rare.

The different variants of tremors could be descriptively classified as rest tremors, postural/kinetic tremors of the type seen in classic ET, and postural/kinetic tremor indistinguishable from enhanced physiologic tremor. However, there are no clear criteria to label them as essential or enhanced physiologic tremor. Therefore, we propose to subdivide them strictly according to the clinical signs and we consider the previously mentioned pathophysiologic assignments as premature.

2.1.5.1 Type I, classic parkinsonian tremor—rest tremor or rest and postural/kinetic tremor with the same frequency

The patient has rest tremor and may or may not have postural/kinetic tremor. The tremor is inhibited during movement and may reoccur during postural tasks. The rest and posture/kinetic tremors have a similar frequency.

Comment: Pure rest tremor does occur in a significant portion of patients. The frequency of pure rest tremor is more than 4 Hz but the upper frequency limit has not been well defined.

Higher rest tremor frequencies of up to 9 Hz can be found, especially in the early stages.[114,115] The clinical observations fit the hypothesis that this form of postural/kinetic tremor (with similar frequencies for both the rest and postural/kinetic tremors) is a continuation of the rest tremor under postural, kinetic conditions, or both. The frequencies for rest and postural/kinetic tremor can be considered to be similar if they do not differ by more than 1.5 Hz. There is frequently a pause in the tremor during the transition from rest to posture.

2.1.5.2. Type II, rest and postural/kinetic tremors of different frequencies

In this case, the postural/kinetic tremor has a higher (>1.5 Hz) and nonharmonically related frequency to the rest tremor.

Comment: A mild form of kinetic tremor is present in almost every parkinsonian patient; it can be detected easily by analyzing slow flexion/extension movements. Sometimes this postural/kinetic tremor can be disabling.

Some patients have a predominant postural tremor in addition to their rest tremor. This form has often been considered to be a combination of an ET with PD[10,114,116] and is rare (<10% of patients with PD) according to our experience. Some of these patients have had postural tremor longer than PD. The critical difference in frequencies is 1.5 Hz. With some experience, this frequency difference can be seen clinically.

2.1.5.3 Type III, pure postural/kinetic tremor

Isolated postural and kinetic tremors do occur in PD. Their frequency may vary between 4 and 9 Hz.

Comment: Postural tremors are common in the akinetic rigid variant of PD. Severe postural and even intention tremor is rare in PD. These tremors have often been considered as ET variants or have been found to be indistinguishable from enhanced physiologic tremor.

2.1.5.4 Monosymptomatic rest tremor

The following criteria characterize a special tremor form that often creates diagnostic problems:

1. Pure or predominant rest tremor.
2. No signs of bradykinesia, rigidity, or problems with stance-stability sufficient to diagnose PD.
3. Duration of the tremor of at least 2 years.

Comment: Some patients exhibit a rest and/or postural tremor without signs of bradykinesia or rigidity significant enough to diagnose PD. Therefore, the clinical findings are not sufficient to diagnose PD although there is positron-emission tomography (PET) evidence that at least some of these patients have a dopaminergic deficit.[117] When such a syndrome persists, we propose to label it provisionally in a descriptive form as monosymptomatic rest tremor. The 2-year period proposed is arbitrary but reasonable according to the natural course of regular PD.[118] The clinical features of this tremor form are for the most part identical with classic parkinsonian tremor.

2.1.6. Cerebellar tremor syndromes

The following conditions have to be fulfilled for the diagnosis of cerebellar (intention) tremor:

1. Pure or dominant intention tremor, uni- or bilateral.
2. Tremor frequency mainly below 5 Hz.
3. Postural tremor may be present, but no rest tremor.

Comment: Cerebellar tremor is often used synonymously with intention tremor although various clinical expressions of tremor have been described in cerebellar disorders.[34,119,120] We have labeled only intention tremor as cerebellar tremor because it has become common practice among clinicians.

Titubation is another tremor that is probably a result of pathology of the cerebellum or its afferent/efferent pathways and is a slow-frequency oscillation depending on postural innervation. The head or trunk can be affected.[120]

Comment: With respect to a tremor differential diagnosis, we refer to intention tremor as the most common form of cerebellar tremor. The tremor resulting from cerebellar abnormalities is often associated with dysmetria, and dyssynergia, hypotonia, or both. All of the other forms of tremor (postural tremor, stance tremor, and so forth) are only accepted as being of cerebellar origin if other signs of cerebellar dysfunction are seen in the patient. With the present definition, a cerebellar tremor can be considered a symptomatic tremor. The major differential diagnoses separate rare forms of ET occurring with predominant intention tremor and some symptomatic tremor etiologies, such as Wilson's disease, and so forth. If the action tremor is severe, rest tremor may occur because the patient is unable to completely relax.

2.1.7. Holmes' tremor

The following criteria apply to this tremor[27,121,122]:

1. Rest and intention tremor with sometimes irregular presentation. In many patients postural tremor is also present. The tremor is often not as rhythmic as other tremors.
2. Slow frequency, usually less than 4.5 Hz.
3. If the time when the lesion occurred can be identified (for example, as in a cerebrovascular accident), a variable delay (usually 4 weeks–2 years) between the lesion and the first occurrence of the tremor is typical.

This is a unique tremor syndrome usually of symptomatic origin resulting from lesions of the central nervous system (CNS). It has been labeled in the past under different names (rubral tremor, midbrain tremor, thalamic tremor, myorhythmia, Benedikt's syndrome).

Comment: The name of this tremor syndrome has long been a matter of debate. The traditional terms rubral tremor and midbrain tremor are considered misleading because more and more cases with lesions outside these classic locations are described with the same or a similar phenomenology.[88,122–132] To avoid names that include topographic descriptions, Holmes' tremor has been accepted in Kiel as the new term, because G. Holmes gave one of the first concise descriptions of this syndrome (although under a different name).[133]

It is generally accepted that this is a symptomatic tremor that can occur from different lesions centered to the brain stem/cerebellum and the thalamus. Two systems, the dopaminergic system and the cerebellothalamic system, must have lesions according to pathoanatomic and PET data.[121,124,129,134] We assume that these systems also have lesions along the further projections of their fiber tracts, thus this tremor might also be caused by lesions in other locations (for example, at multiple cortical sites).

One open question is whether thalamic tremor[122,123,127,131,135] is a separate entity or only a variant location of the underlying pathology. At present, we propose to label the clinical syndrome as Holmes' tremor and the etiology as tremor after a midbrain/thalamic lesion. Most of the patients with so-called thalamic tremor have some additional dystonia and classic signs indicating a thalamic lesion. Some patients with Holmes' tremor may have additional dystonia, which raises the question of whether their condition should be classified within the dystonic tremor syndromes.

2.1.8. Palatal tremor syndrome

Palatal tremor can be separated into two forms.[136,137] *Symptomatic palatal tremor* is characterized by:

1. Preceding brain stem/cerebellum lesion with subsequent olivary hypertrophy, which can be demonstrated with magnetic resonance imaging (MRI) scans.
2. Rhythmic movements of the soft palate (levator veli palatini) and often other brain stem-innervated or extremity muscles.

Essential palatal tremor is characterized by:

1. Absent preceding lesions and absent olivary pseudohypertrophy.
2. The patient usually has an ear click. The rhythmic movements of the soft palate mainly involve the tensor veli palatini. Extremity or eye muscles are not involved.

Comment: Besides these two typical forms of palatal tremor, further rhythmic movement disorders of the palate and neighboring structures may occur that cannot yet be classified. The name, palatal myoclonus, has been omitted because pathophysiologic and pathoanatomic data favor use of the term tremor; some use the term segmental cranial tremor. Psychogenic palatal tremor has been observed. Other rhythmic movement disorders of cranial nerve muscles to be mentioned are the rabbit

TABLE 5. *Etiologic classification of tremor*[27]

Disease	Classification
1. Hereditary, degenerative and idiopathic diseases	
Parkinson's disease	R, P
Juvenile Parkinson's disease	R, P
Pallidonigral degeneration	P
Multiple system atrophy	
Olivopontocerebellar atrophy	R, P, I
Striatonigral degeneration	R, P, I
Wilson's disease	R, P, I
Progressive pallidal atrophy	R
Huntington's disease	R, P, I
Benign hereditary chorea	P, I
Fahr's disease	R, I
Paroxysmal dystonic choreoathetosis	I
Familial intention tremor and lipofuscinosis	I
Ramsay-Hunt syndrome (progressive myoclonic ataxia)	P, I
Ataxia telangiectasia	P
Dystonia musculorum deformans	P
DOPA-responsive dystonia	R, P
Spasmodic torticollis	P
Meige syndrome	P
Essential myoclonus and tremor	P, I
Essential tremor	R, P, I
Task-specific tremors	
Writers tremor	
Voice tremor	
"Golfers"-tremor ("yips")	
Laughing tremor	
Klinefelter's syndrome	P, I
2. Cerebral diseases of various etiologies	
Infectious diseases and other inflammations	
Multiple sclerosis	R, P, I
Neurosyphilis	R, P, I
Neuroborreliosis	R, P, I
HIV	P
Smallpox, measles	P
Typhus	P
Space-occupying lesions	
Tumors	R, P, I
Cysts	R, P, I
Hematoma	R, P, I
A/V malformations	R, P, I
Cerebrovascular insults	R, P, I
Trauma	R, P, I
3. Metabolic diseases	
Hyperthyroidism	P
Hyperparathyroidism	R, P
Hypomagnesemia	R, P
Hypocalcemia	R, I
Hyponatremia	P
Hypoglycemia	P
Chronic hepatocerebral degeneration	P, I
Hepatic encephalopathy	P, I
Kidney disturbances	P, I
Lack of vitamin B12	R, P, I
Eosinophilia myalgia syndrome	P, I
4. Peripheral neuropathies	
Charcot-Marie-Tooth	P, I
Roussy-Levy syndrome	P, I
Chronic demyelinating neuropathies	P, I
Guillain-Barré syndrome	P, I
Gammopathies (IgM, IgG)	P
Malabsorption neuropathy	P
Reflex sympathetic dystrophy	P
Polyneuropathy of various origins (diabetes, uremia, porphyria)	P
Spinal muscular atrophy	P
Kennedy's syndrome	P, I
Nicotine	P
Mercury	P, (R, I)
Lead	P, (R, I)
CO	P, (R, I)
Manganese	P, (R, I)
Arsenic	P, (R, I)
Cyanide	P
Naphthalene	P, I
Alcohol	P, I
Toluene	P
DDT	P, (R, I)
Lindan	P, I
Kepone	P, I
Dioxins	P
6. Drugs	
Centrally acting substances	
Neuroleptics	R, P
Reserpine	R, P
Tetrabenazine	R, P
Metoclopramide	R, P
Antidepressants (especially tricyclics)	P
Lithium	R, P, I
Cocaine	P
Alcohol	P, I
Sympathomimetics (for example, adrenaline)	P, I
Bronchodilators (β_2-agonists)	P, I
Theophylline	P
Caffeine	P
Dopamine	P
Steroids	
Progesterone (medroxyprogesteronactate)	R, P
Antiestrogenes (tamoxifen)	P
Adrenocorticosteroids	P
Miscellaneous	
Valproate	P
Perhexiline	R, P
Antiarrhythmics (amiodarone)	P
Mexiletine, procainamide	P
Calcitonin	P
Thyroid hormones	P
Cytostatics (vincristine, adriablastine, cytosinarabinoside, ifosfamide)	P
Immunosuppressants (cyclosporin A)	P, I
7. Others	P
Emotions (anxiety, stress)	P
Fatigue	P
Cooling	P
Trauma of the periphery/reflex sympathetic dystrophy	P, I
Withdrawal of drugs	P
Withdrawal of alcohol	P, I
Withdrawal of cocaine	R, P
Psychogenic tremor	R, P, I

R, rest tremor; P, postural tremor; I, intention tremor.

syndrome, a medium-frequency tremor of lip muscles and geniospasm (see section 2.1.3.4.3).

2.1.9. Drug-induced and toxic tremor syndromes

A tremor is considered to be drug-induced if it occurs in a reasonable time-frame following drug ingestion. Toxic tremors occur after intoxication.

Comment: Drug-induced tremors can have the whole range of clinical features of tremors. The clinical presentation depends on the drug and possibly (so far undefined) individual dispositions of the patients may play a significant role (for example, unusual tremor syndromes in patients with ET). The most common form is the enhanced physiologic tremor syndrome that occurs after use of sympathomimetics or antidepressants. Another frequent form is classic parkinsonian tremor that follows use of neuroleptic or dopamine-receptor blocking drugs. Cerebellar tremor syndromes may occur after intoxication by lithium and other substances.

The tremor caused by withdrawal from alcohol or other drugs has been characterized as enhanced physiologic tremor with tremor frequencies above 6 Hz.[138–145] However, this condition has to be separated from the intention tremor of chronic alcoholism that is likely to be related to cerebellar damage after alcohol ingestion.[138] This is often associated with a 3-Hz stance tremor that has been ascribed to the anterior lobe damage caused by chronic alcoholism.[146] The different etiologies of drug-induced and toxic tremors are listed in Table 5.

A specific variant is *tardive tremor* after long-term neuroleptic treatment.[147–149] It occurs with a frequency range of 3–5 Hz, and is most prominent during posture but is also present at rest and during goal-directed movements. The rabbit syndrome is also considered to be a form of tardive tremor.[150]

The diagnosis of toxic tremors includes other clinical signs of CNS intoxication (for example, eye movement abnormalities, gait disturbance).

2.1.10. Tremor syndromes in peripheral neuropathy

This condition is assumed if a tremor develops in a patient in association with a peripheral neuropathy.

Comment: Tremors develop in patients with some forms of peripheral neuropathy more often than others. Many neuropathies have been found to be associated with tremor[151–163] (see Table 5) but demyelinating neuropathies, and especially dysgammaglobulinemic neuropathies, are frequent causes of such tremors.[152,164] The tremors are mostly postural and kinetic tremors. The frequency in hand muscles can be lower than in proximal arm muscles in patients with gammopathies. It should be mentioned that abnormal position sense is not a required condition for the diagnosis. The pathophysiology of this tremor is thought to result from the abnormal interaction of peripheral and central factors.[152]

The coexistence of peripheral neuropathy and ET is sometimes present and it may be impossible to distinguish the etiology in a particular patient.

2.1.11. Psychogenic tremor

Psychogenic tremors may have different clinical presentations. The following criteria suggest psychogenic tremor[165,166]:

1. Sudden onset of the condition, remissions, or both.
2. Unusual clinical combinations of rest and postural/intention tremors.
3. Decrease of tremor amplitude during distraction.
4. Variation of tremor frequency during distraction or during voluntary movements of the contralateral hand.
5. Coactivation sign of psychogenic tremor.
6. Somatization in the past history.
7. Appearance of additional and unrelated neurologic signs.

Comment: The diagnosis of psychogenic tremor is not only a diagnosis of exclusion but a positive one based on the anamnestic data and clinical findings mentioned earlier. Clinical findings 2–5 are usually positive in most patients with psychogenic tremors.

2.1.12. Unclassified tremors

If a tremor cannot be classified, it should be described with the terms defined in "Phenomenology of Tremor" and labeled as unclassified.

2.2. Etiologic classification

Etiologic classifications of tremor are never complete and need to be continuously updated. A list of etiologies, including the described clinical features is included (see Table 5).

3. DIFFERENTIAL DIAGNOSIS

The following syndromes may be misinterpreted as tremors.

3.1 Rhythmic myoclonus and so-called cortical tremor

Rhythmic myoclonus is a syndrome that has been proposed by several authors with the following definitions:

1. Intermittent, brief muscle jerks, irregular or rhythmic, arising in the CNS.
2. Slow frequency (usually below 5 Hz).
3. Hyperkinesia usually limited to segmental levels.

Comment: This type of hyperkinesia has not yet been well defined and the present proposal is preliminary.[10,167–169] Rhythmic myoclonus cannot be clearly distinguished from tremor (especially from Holmes' tremor and its different variants). Sometimes the driving muscle contractions are brisk so that there are longer pauses between the individual jerks; this has been put forward as a feature for the differential diagnosis.[167] The present definition also does not separate epilepsia partialis continua from rhythmic myoclonus.

Cortical tremor is not a tremor but a specific form of rhythmic myoclonus[170,171] that has also been described as an inherited form.[172] It consists of:

1. High-frequency, irregular tremor-like postural and kinetic myoclonus almost indistinguishable from high-frequency postural tremor.
2. Synchronous, short, high-frequency jerks (7–18 Hz) on an EMG.

Comment: On electrophysiologic work-up these patients show the typical features of cortical myoclonus with a related electroencephalogram (EEG) spike preceding the EMG jerks and often enhanced, long-loop reflexes, giant somatosensory evoked potentials (SEPs), or both. This form of myoclonus is seen in different conditions without specific diagnosis[170,171] in familial cortical myoclonic tremor[172] but also in corticobasal degeneration[173–176] and Angelman's syndrome.[177]

It must be emphasized that so-called jerky tremors are often not tremors but rather, myoclonus or asterixis. Therefore, it is recommended that additional electrophysiologic tests be performed for differential diagnosis of these conditions (polymyography, median nerve SEPs, long-latency reflexes, EEG, and jerk-locked averaging).

Spinal myoclonus can be rhythmic at a fast rate[167] and probably should be considered a separate entity.

3.2 Asterixis

Asterixis is a negative myoclonus with sudden lapses of innervation. When the EMG pauses are long (>200 ms), typical flapping tremor during tonic contraction results. When the pauses are shorter, the clinical phenomenology resembles a somewhat-irregular, high-frequency tremor.

Comment: Asterixis can occur either as a focal or a generalized condition and is usually a symptomatic movement disorder. Unilateral asterixis is often caused by focal lesions of the contralateral hemisphere and bilateral asterixis commonly results from metabolic or endocrine dysfunction, intoxication, and focal brain stem lesions.[169] The diagnosis can sometimes be suggested by visible, sudden lapses of the outstretched hand, but it is often necessary to confirm the diagnosis with polymyographic recordings from different muscles of one extremity showing synchronous pauses of activation.[169]

3.3 Clonus

Clonus is a rhythmic movement mainly around a joint (but sometimes of a whole extremity) elicited through the stretch reflex loop and increasing in strength (or amplitude) by maneuvers affecting the stretch reflex.

Comment: Clonus is only rarely misinterpreted as tremor.[178] On clinical examination, passive stretching of the muscles increases the force of clonus but not of tremor. This is the best clinical criterion separating the two conditions. In terms of pathophysiology, the question of whether clonus is dependent on a central oscillator[168,179] or only based on segmental reflex circuits is still being debated.

3.4 Epilepsia partialis continua

Epilepsia partialis continua (EPC) is a focal epilepsy that can produce (mainly low-frequency) rhythmic jerks of an extremity and thus can be misinterpreted as tremor.

Comment: Rest (and rarely postural/intention) tremors can be mimicked by EPC. A lack of tremor history, a medical history of epilepsy, and presence of EEG spikes, short EMG bursts, and jerk-locked averaging are helpful to identify this movement disorder. It is one of the more common misdiagnoses of Holmes' tremor.

3.5 Rhythmic dyskinesias and stereotypies

Levodopa-induced diphasic dyskinesias are often repetitive and have a slow rhythm (1–3 Hz). Although they may appear as tremor-like, they are not included here as tremors. Stereotypies may have rhythmic components but nevertheless are dominated by more complex movements.

4. USEFUL TECHNICAL INVESTIGATIONS FOR THE DIAGNOSIS AND MONITORING OF TREMORS

4.1. Electromyography of tremors

The diagnosis of tremors can be supported by EMG examinations; a surface EMG is usually sufficient and needle recordings are rarely necessary. This is the best method for identifying those muscles and limb segments involved in tremor.[25] An EMG assessment of tremors is the most reliable method of diagnosing primary orthostatic tremor[62,64,66] and of confirming or excluding asterixis[61,169] in cases of high-frequency, irregular tremors. It can also be necessary for confirming laryngeal tremor.

An EMG examination can sometimes be helpful in the diagnosis of dystonic tremor and for assessing tremor frequency.[82,103,105,106] The value of EMG examination is limited for differentiating ET from parkinsonian tremors.[110,113,180–183] However, it is often necessary for targeting botulinum toxin treatment.

4.2. Fourier analysis of tremor

Use of time series analysis for tremor has been well established,[25] although some methodologic concerns need to be taken into account.[184,185] Relevant information obtained from such analyses are the frequency and the amplitude of tremor curves. For some tremors, like Holmes' tremor and primary orthostatic tremor, the frequency is a most valuable diagnostic tool.[61]

4.3. Advanced methods of time series analysis

Nonlinear time series analysis (like chaos analysis) is just beginning to be applied to tremor time series[186,187] and has not yet led to substantial results. More simple classifications referring to specific nonlinear properties of tremor time series may be helpful in separating essential and parkinsonian tremor curves.[188,189] Therefore, we consider these techniques to be predominantly experimental procedures that do not as yet have a routine clinical application.

4.4. Three-dimensional analysis of tremor curves

Although the study of three-dimensional movements seems to be promising, little work has been done in this field.[190,191] It could be used to quantify the envelope of tremor curves.

4.5. Graphic tablet analysis

Graphic tablets have been introduced to analyze writing tremor and to measure the extent of kinetic tremor,[182,192] which can occasionally be used for diagnostic purposes and especially for treatment monitoring.

4.6. Long-term recordings of tremor

Long-term recordings of EMG have been assessed for validity and reliability.[193–195] The technique is particularly useful for measuring tremor occurrence rate (the proportion of time that tremor is present). It may be useful for studying the response of tremor to treatment (for rest tremors in particular).

4.7. Reflex testing

Although reflex testing has been used in several studies to distinguish different conditions,[168,196–198] it has not been accepted to differentiate different forms of tremor. Electrically elicited hand muscle reflexes are necessary for the differential diagnosis of rhythmic myoclonias presenting as jerky tremors.[173,174,199,200]

4.8. Activation patterns of ballistic movements

This means of investigating tremor for scientific purposes[87,201] has not been used for diagnoses or for treatment monitoring.

4.9. Single motor unit recording

This technique allows interesting insights into the mechanisms of tremor.[202–207] Hitherto, no clear-cut diagnostic or treatment-related applications have been described.

4.10 Median nerve SEPs, jerk-locked averaging, and EEGs

The differential diagnosis of myoclonus and tremor may be impossible on the basis of clinical investigation alone. For this purpose, these classic electrophysiologic techniques may be necessary.[199,200]

5. TREMOR STUDY DESIGN

The primary purpose of this statement is to provide a uniform nomenclature for the classification of tremor and thus it was decided that the complex issues concerning treatment of the different tremor syndromes should not be discussed in detail. However, the purpose of the document is to allow clinical research to be undertaken on patient populations that are clearly defined to the satisfaction of the scientific community. Consequently, we also recommend certain standards for therapeutic trials and specifically, that drug trials have a randomized double-blind design and include placebo control wherever appropriate.

For scientific experiments, specific neurophysiologic tests may be sufficient, but for assessing the value of a specific medical therapy, a more comprehensive test battery is advocated, including:

1. A clinical rating scale for tremor impairment (objective tremor scale).
2. Measures of performance (peg board test, volumetric methods, and so forth).
3. A disability scale.
4. An assessment of handicap and quality of life.
5. A neurophysiologic measure (accelerometry, EMG, and so forth).

LEGENDS TO THE VIDEOTAPE

These videotaped case examples were provided by P. Bain (London, England); M. Brin (New York, New York, U.S.A.); G. Deuschl and J. Raethjen (Kiel, Germany); and H. Shibasaki (Kyoto, Japan). The videotape

was produced by J. Raethjen and F.W. Lehmhaus at the Institut für Pädagogik der Universität Kiel.

Case 1: Classic ET (Dr. Bain). A 67-year-old man with a 10-year history of action tremor of the hands; the tremor is alcohol and propranolol responsive. There is a family history of tremor affecting at least three generations. The videotape segments show the patient's upper limb postural and kinetic tremor, which is evident during the finger-to-nose test.

Case 2: ET with intention tremor (Dr. Bain). A 68-year-old woman with a long-standing history of head and voice tremor. She has been suffering from additional hand tremor during action, especially goal-directed movements, for 8 years. She has a family history that is positive for tremor but no response to alcohol. The voice and head tremor are shown in the first part of the video; the intention tremor with the typical amplitude variation can be seen in the finger-to-nose test.

Case 3: Orthostatic tremor (Drs. Deuschl and Raethjen). This woman had unsteadiness of stance for 10 years. She has no problems during walking and sitting and is able to walk for hours. There is no tremor of the hands or head. On clinical examination, the only finding is a slight shivering of the quadriceps muscle that can hardly be seen but can be palpated. The definite proof of orthostatic tremor can be accomplished by an EMG recording of a 16-Hz pattern of the quadriceps muscle during standing.

Case 4: Voice tremor (Dr. Brin). A 47-year-old woman with a 10-year history of tremor of the voice. Tremor of the hands is very slight in the left hand and is only present during goal-directed movements. The video illustrates the differing tremor amplitudes for different sounds, notes, and volume of the voice.

Case 5: Dystonic tremor I (Dr. Bain). A 33-year-old woman with a 12-year history of spasmodic torticollis and head tremor that developed after a mild head injury; generalized dystonia and tremor of the arms and legs subsequently developed. The videotape shows the patient's marked tremulous torticollis and coarse dystonic action tremor of the arms and legs. The head tremor and dystonia responded partially to treatment with botulinum toxin.

Case 6: Dystonic tremor II (Dr. Bain). Pain and tremor developed in this 58-year-old, right-handed woman in the right hand on writing at the age of 30. Five years later a postural tremor appeared in her right hand. She switched to writing with her left hand but over the next decade tremor developed in her left hand and the postural tremor in the right arm increased in severity. The videotape segment demonstrates tremor and abnormal posturing of the left hand on writing and a postural tremor of the right hand that is exacerbated when she writes.

Case 7: Dystonic tremor III (Drs. Deuschl and Raethjen). A 38-year-old woman who has had a postural and writing tremor of her right hand for approximately 2 years. The video shows the typical low-frequency tremor with a variable amplitude. In the second part of the video, the tremor is reduced markedly by full relaxation in a supine position. As soon as the position of the hand/arm requires minimal muscle activation, the tremor amplitude increases again.

Case 8: Primary writing tremor (Drs. Deuschl and Raethjen). A 44-year-old man with a 4-year history of a slowly progressive abnormality during writing that he found very disabling lately in his job as a teacher. There is no family history and no effect of alcohol on the tremor. There is no tremor under rest, postural, and kinetic conditions; however, during writing he has an obvious, high-amplitude pronation–supination tremor. The writing tremor was subsequently reduced markedly by botulinum toxin injections into the pronator quadratus muscle in the right forearm.

Case 9: Position/task-specific tremor (Dr. Bain). This 34-year-old man is a professional drummer with a 4-year history of tremor in the left hand that appears when he plays the drums. It is worse when he uses the traditional rather than the matched grip. There is no significant family history. The videotape shows that the tremor is released when the hand is semipronated and becomes more pronounced when the left thumb exerts downward pressure on the drumstick. The tremor responds partially to alcohol and propranolol.

Case 10: PD: classic pure rest tremor (Drs. Deuschl and Raethjen). A 67-year-old man with a 2-year history of PD with mild, left-sided rigidity and akinesia. His main complaint was that his left-sided hand tremor occurred only under resting conditions.

Case 11: PD, combination of rest and postural/action tremor (Drs. Deuschl and Raethjen). A 50-year-old woman with PD who had postural tremor of the left hand for 5 years before the symptoms of akinesia, rigidity, and rest tremor developed. The videotape shows that she now has a bilateral rest and postural tremor of the same frequency.

Case 12: PD, combination of rest and higher-frequency postural/action tremor (Drs. Deuschl and Raethjen). A 55-year-old man with a 10-year history of PD. Tremor was his major complaint during the first years; now his main problem is motor fluctuations. Tremor occurs during on-and-off phases. The interesting feature of this case is the difference in frequency between rest and action tremor. Although the rest tremor fre-

quency is 5 Hz, the postural and kinetic tremors occur at 7.3 Hz, which is illustrated in the power spectra of accelerometric tremor recordings under the different conditions.

Case 13: Monosymptomatic rest tremor (Drs. Deuschl and Raethjen). A 49-year-old man with a 3-year history of rest tremor in the right hand with a mild postural component of the same frequency. There were no signs or symptoms of akinesia or rigidity. However, a recent PET study in this patient demonstrated a reduced fluorodopa uptake in the striatum, and we thus assume that this condition is a variant of PD.

Case 14: Cerebellar tremor, classic intention tremor (Drs. Deuschl and Raethjen). A 28-year-old woman who was diagnosed with multiple sclerosis (MS) 4 years ago. Her presenting complaints were a marked intention tremor of her right hand and gait ataxia. The typical amplitude fluctuations of intention tremor can be seen during the finger-to-nose test.

Case 15: Cerebellar tremor, titubation (Dr. Bain). A 48-year-old man with a 4-year history of clinically definite MS that was diagnosed shortly after a head injury. The videotape shows his head titubation, which is improved when the patient's forehead is lightly touched.

Case 16: Holmes' tremor, midbrain lesion (Drs. Deuschl and Raethjen). A 56-year-old man who had a brain stem trauma. Holmes' tremor developed 10 years ago and shows the typical combination of rest and intention tremor at a low frequency in his right hand; he also has some postural tremor. At the time of the recording of this videotaped segment, the patient was treated with anticholinergics and levodopa, which had a good effect on tremor severity. (An earlier videotaped segment of this patient has already been published in *Movement Disorders.*[121])

Cases 17 and 18: Essential and symptomatic palatal tremor (Drs. Deuschl and Raethjen). These two cases have already been published in the *Movement Disorders* videotape series. They illustrate the different palatal movements in essential and symptomatic palatal tremor. In the case of essential palatal tremor, the roof of the soft palate is elevated because of activation of the tensor veli palatini muscle. In the case of symptomatic palatal tremor, the edge of the soft palate is moving as a result of activation of the levator veli palatini muscle.

Case 19: Psychogenic tremor (Drs. Deuschl and Raethjen). A 41-year-old woman who had a rest, postural, and action tremor of the right hand in combination with muscle pain in the right shoulder girdle for 2 years. During the examination there is no consistent activation pattern of the tremor. In addition her hand tremor develops, usually in combination with a slight co-contraction, leading to a constant tiny dorsal extension of the hand. Under changing loads, the tremor amplitude often shows fluctuations in these patients, which is illustrated by a mild increase in tremor amplitude on loading the hand and a marked deterioration of tremor when the load is taken away.

Case 20: Tremor in gammopathy (Drs. Deuschl and Raethjen). A 64-year-old man who had gammopathy for 15 years. His postural and action tremor was slowly progressing during the last 2 years. The videotape demonstrates a postural tremor, mainly in the fingers, and a more marked action and kinetic tremor when he is pouring water from the glass. The tremor shows the typical irregularities with varying tremor amplitudes.

Case 21: Tardive tremor (Dr Brin). A 44-year-old woman who was placed on highly potent neuroleptics 6 years ago. Involuntary, rhythmic movements of the tongue and jaw developed 1 year ago when she was still on 6 mg Haldol per day. This movement disorder shows some typical features of a tardive tremor.

Case 22: Rhythmic cortical myoclonus (Dr. Shibasaki). A 74-old-woman with slowly progressive or almost-stationary hand tremor of the postural as well as the action type since age 40. He had no history of generalized convulsions. Her 38-year-old son has had attacks of unconsciousness and hand tremor since the age of 35. Electrophysiologic findings fulfill the criteria for cortical reflex myoclonus. Beta blocker was not effective but she responded to anticonvulsants. The final diagnosis was familial cortical myoclonic tremor.

Case 23: Rhythmic spinal myoclonus (Dr. Shibasaki). A 19-year-old woman with intermittent and slowly progressive rhythmic movements of high frequency in the legs. There was no family history or consanguity. The diagnosis of spinal myoclonus of unknown etiology was made.

Case 24: Asterixis (Dr. Shibasaki). A 20-year-old man with generalized convulsive seizures, myoclonic jerks, and slowly progressive mental deterioration since the age of 15. The diagnosis of Lafora disease was made by skin biopsy. The family history contains information positive for this condition. The patient shows negative myoclonus of the hand muscles that are also elicited by electrical stimulation of the median nerve.

Case 25: Asterixis (Dr. Shibasaki). Typical asterixis or negative myoclonus of subcortical origin in a patient with hepatic encephalopathy.

Case 26: Epilepsia partialis continua (Dr. Shibasaki). This 10-year-old girl had a lesion that appeared on MRI in the right parietal lobe, manifesting as rhythmic jerks in the right foot.

APPENDIX

Videotaped recording of patients for diagnostic purposes:

To standardize the clinical presentation of patients, the following sequences are proposed for video recordings.

Rest tremor:

Patient sitting, arms relaxed on arms of a chair. Patient is asked to count backward from 100. When the tremor is clearly visible, the patient is asked to lift his or her hands, to hold them outstretched and make a finger-to-finger test.

If the patient cannot relax for any reason and there is the suspicion of residual postural innervation of the arms, the patient should lie on a bed and count backward in this position.

Postural tremor:

Patient standing or sitting, holding arms outstretched first and then in shoulder abduction, with flexed elbows and the fingers near the head but without visual feedback.

Simple kinetic tremor:

Tested during slow flexion/extension movements of the hands or any other nonvisually guided and nondirected movement.

Intention tremor:

Finger-to-finger test, if possible finger-to-nose test.

Additional performance tests:

- Writing a sentence.
- Drawing a spiral.
- Pouring water from one glass into another.

The topography of the tremor in a particular patient should be documented by short sequences (for example, chin, jaw, voice, legs). If a particular region is not affected, it does not have to be filmed. Special aspects not mentioned here (for example, position-specific tremors) should be documented in the most appropriate form.

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